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THORACIC ECTOPIA CORDIS - A CASE REPORT

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ABSTRACT

Ectopia Cordis is a rare congenital anomaly characterized by partial or complete displacement of the heart, outside the thoracic cavity. Usually ectopia cordis is associated with other multiple anomalies and intra cardiac defects ,due to ventral body wall developmental defects.

The present case was a fullterm 6 hours old female baby, weighting 1.8 kg ,born to a 22 year old, 2nd Gravida, para2 mother, delivered by normal vaginal delivery. No history of consanguineous marriage. Baby has ectopia cordis, complete bilateral cleft lip and cleft palate and sternum is absent. After investigations single staged surgery was performed. The baby died after three hours due to cardiac and respiratory arrest.

Keywords: Ectopia cordis, developmental anomaly, ventral body wall developmental defects, absence of sternum.

1. INTRODUCTION

Haller[5] first described the term Ectopia cordis in 1706.The term was derived from Greek word *ektopos* meaning away from a place. Ectopia cordis is defined as complete or partial displacement of the heart outside the thoracic cavity. Ectopia cordis may occur as an isolated malformation or it may be associated with a large category of ventral wall defects that effect the thorax, abdomen or both. All these are due to defects in ventral body wall mid line fusion during development. The incidence of Ectopia cordis is 5.5-7.9 in 1 million births. More in females.

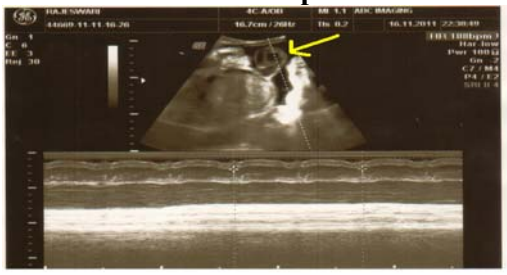
Kim[7] *et al*, in 1997 classify the Ectopia cordis into 5 types. Depending up on the position of the heart they are 1.Thoracic, 2.Abdominal, 3.Thoraco-abdominal, 4.Cervical,and cervicothoracic. Thoracic type is more common and cervical type is rarest. The thoraco-abdominl Ectopia cordis associated with sterna cleft, diaphragmatic hernia, Omphalocele, and an intracardiac defect is known as Cantrell's pentalogy.

2. CASE REPORT

A full term 6 hour old, female baby, born to a 22years ,Gravida 2, para 2 ,mother by normal vaginal delivery at Government Hospital on 26-11-2011,that is 90 km from our place. Then baby was shifted to Government General Hospital, Guntur. At the time of birth APGAR at 1 minute was 8, weight was 1.8 kg. Heart rate is 170/minute; respiratory rate is 54/minute. Cry is feeble. Heart sounds s_1, s_2 present, no murmurs are present. No organomegaly. Mother underwent antenatal checkups at local Doctor. There was no history of consanguineous marriage, no family history of congenital anomalies, no history of intake of teratogens, no history of exposure to radiation. No Rh incompatibility. No history of vaccination against Rubella.2 doses of T.T was given. Mother is not an alcoholic.

Antenatal ultrasonography has been done at local Doctor, but the deformity was not detected, probably due to lack of experience.

Fig. No.1. Ultrasonography during antenatal period



Arrow showing the presence of heart outside the thoracic cavity.

On examination baby had pulsating heart covered with a serous membrane was present in the mid line in pectoral region of thoracic wall. Along with Ectopia cordis she had complete cleft lip and cleft palate, sternum is absent, and divarication of recti muscles are present. Umbilical cord normal.

Fig. No.2. Baby with Thoracic Ectopia cordis Showing naked heart lying outside the thoracic cavity.

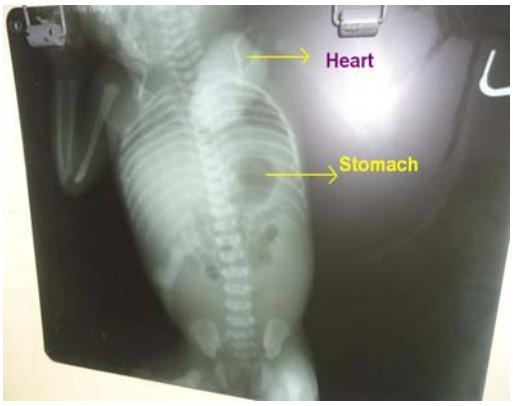
Fig. No. 3.Thoracic Ectopia cordias with cleft lip and normal umbilical cord

No neck swelling, no limb abnormalities are found.

Investigations: All routine blood tests are normal . Other investigations are

a. X-ray chest.

Fig. No.4. Sternum is absent. Ribs are crowded .stomach pushed into chest wall.



b. Ultrasonography : Abdomen normal.

Fig. No.5

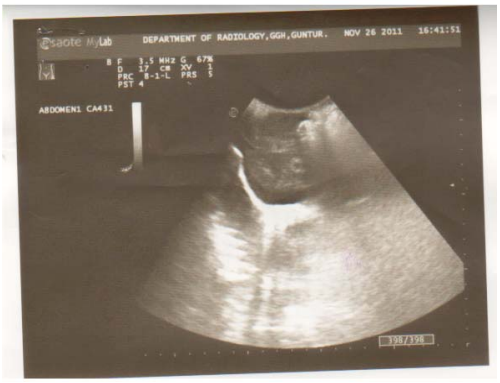
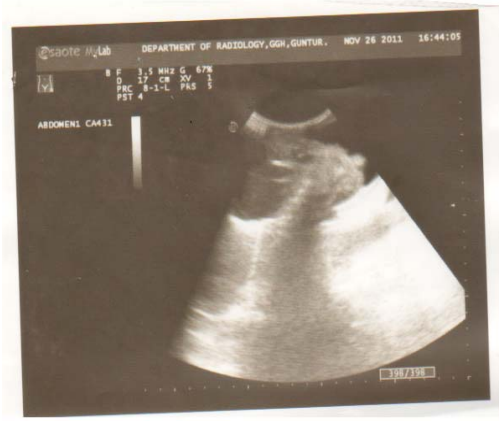


Fig. No.6.



c. **2D Echo: Findings are :** Single atrium, ostium primium ASD,two vantricles are present, inter ventricular septum intact,left vantricle normal,pericardium normal,E.F.70%.

d. **Neurosonogram: Normal.**

Operation: After investigations Operation was performed. Single stege operation has ferformed.After covering the heart with skin,Silo pouch created around the heart and sutured. During the operation presence of diaphragematic herniais was found.Post-operatively baby died after 2 hours due to cardiac and respiratory arrest.

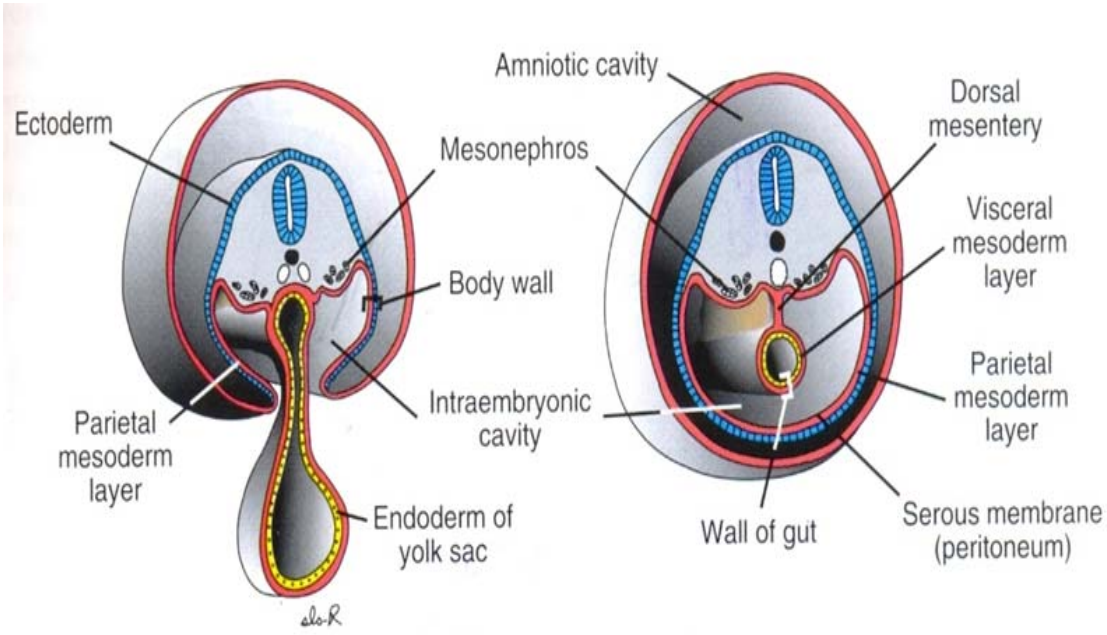
3. DISCUSSION

Ectopia cordis is a rare and stricking congenital heart defect, which was first observed 5000 years ago.[10]. The term ectopia cordis was first coined by Haller[5] in 1706. The defect is described as malposition of heart, partially or completely outside the thorax. It is a rare congenital defect in the fusion of ventral chest wall resulting in extra thoracic location of the heart. Ectopia cordis is usually associated with other congenital anomalies and intra cardiac defects. Depending upon the position of the heart ectopia cordis can be classified into 5 types.

1. Cervical : In which the heart is located in the neck with sternum ie. Usually intact.
2. Thoracocervical : in which the heart is partially in the cervical region but the upper portion of the sternum is split.
3. Thoracic: in which the sternum is split or absent and heart lies partially or completely outside the thorax.
4. Thoracoabdominal : which usually acompanies Cantrell's syndrome.
5. Abdominal : in which the heart passes through a defect in the diaphragm to enter the abdominal cavity. Kim[7] *et al*, 1997; Dobell[4] *et al* 1982.

Pathogenesis: Embrologically ecotopia cordis is due to defect in the closure of lateral body wall folds to form ventral body wall. At the end of third week of embryo the lateral plate mesoderm splits into outer somatic and inner splanchic layers. The space created between the layers of lateral plate mesoderm consitutes the primitive body cavity.

Fig. No.7. Lateral folds of embryo



With courtesy from T.W.SadlerLangman's[9] Medical Embryology. In-Cardiovascular system. 11th edition.
During 4th week the sides of embryo begins to grow ventrally forming two lateral body wall folds. By the end of 4th the lateral body wall folds meet in the mid line and fuse to close the ventral body wall. This closure is aided by head and tail folds. The developed heart reaches the original position by the end of 4th week. Complete or incomplete failure of midline fusion and defect in mesoderm fusion in embryonic state can result in a variety of disorders ranging from isolated ectopia cordis to complete ventral evisceration.

The other organs generally involved in ectopia cordis are

- CNS – Cranial cleft,
  Hydrocephalus,
  Cephalocoele.
- Face - Cleft lip and cleft palate.
- **Cardiac**
  AVSD,
  VSD,
  Tetralogy of Fallot,
  pulmonary stenosis,
  mitral atresia,
  tricuspid stenosis,
  coarctation of the aorta
- **Pulmonary -**
  Hypoplastic lung**.**
- **Skeleta**l-
  scoliosis.
  limb Hypoplasia,
  Syndactyly.
  sternal cleft
- **Abdominal-**
  Divarication of recti muscle,
  Omphalocele,
  diaphragmatic hernia.

Ectopia cordis is frequently associated with Turners syndrome. Carmi[3] *et al* 1993 also supported that these ventral midline defects should be linked up by a X-linked mutation.

Management: Staged surgical procedure is the only choice of treatment.

Prognosis: is generally bad and most cases results in still birth or die shortly after birth due to infection, cardiac failure, or hypoxemia because it is associated with intracardiac and other congenital anomalies.
Surgical correction of Ectopoa cordis is complex and generally requires a staged closure.

1. Covering the heart by skin.
2. Placement of the heart into thoracic cavity and correction of heart defects.
3. Sterna or thoracic reconstruction.

Prevention

- Early diagnosis by routine prenatal Ultrasonography as early as 10-12 weeks of pregnancy.
- And termination of pregnancy prior to viability should be considered after discussion with parents.
- This case is also highlights the need to strengthen the existing referral system and rise the awareness of the Health personal on the correct diagnosis and prompt referral of unusual cases like this.

But there are interesting cases reported in the past they are as follows.
In 1995 Amato[1] *et al* reported successful single stage repair of thoracic E.C.
Christopher Wall 19th Aug 1975 hold the Guinness World Record for the oldest living person with E.C.
Doctors at AIIMS, New Delhi, India successfully operated on a 10th day old male with E.C on 2nd sep 2009. However baby

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